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Relationship Between Early Feeding and Communication Development in Infants: Birth To 12 Months

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This conference paper was originally published as:

Massey, S. J., Hird, K., & Simmer, K. (2004). Relationship Between Early Feeding and Communication Development in Infants: Birth To 12 Months. *2nd World Congress of Pediatric Gastroenterology, Hepatology and Nutrition*.

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Abstracts Submitted Late for the 2nd World Congress of Pediatric Gastroenterology, Hepatology and Nutrition

Please see Volume 39, Supplement 1

P1193

SERUM LEPTIN IN CHILDREN WITH INSULIN DEPENDANT DIABETES IN RELATION TO OBISITY

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Introduction: leptin is an obese gene product, an adipocyte derived hormone that signals the state of energy stores to the brain acting through specific receptors in the hypothalamus(1). leptin plays a crucial role in metabolic homeostasis by reducing feeding behavior(2,3).to investigate wheather leptin concentratio in children with type 1 diabetes was related to metaolic control, body mass index or insulin, serum leptin, serum insulin and insulin-like growth factor-1 levels were estimated in 30 diabetic non obese children and 15 diabetic obese children and 15 lean children as control.

Methods: serum leptin, serum insulin and insulin like growth factor-1 were estimated by ELISE technique. Anthropometric measures,were taken, boy mass index was calculated.

Results: serum of diabetic children either obese or non obese had significantly higher levels of leptin and insulin copared with lean control (P<0.001)and p<0,005 respectively).serum leptin level was significantly correlated with body mass index (BMI) for the combined group (dabetic obese and non obese and lean, r=.055, p<0.01).

Table 1: Charactristics of IDDM non obese, IDDM obese and lean children

	IDDM non obese no.=30 mean=8.43 SD=1.59	IDDM obese no.=15 mean=8.27 SD=1.83	lean control no.=15 mean=7.27 SD=1.87
age in years			
sex (M:F)	16:14	8:7	6:9
height (cm)	mean=129.27 SD=9.38	mean=124.67 SD=10.55	mean=117.33 SD=9
weight (Kg)	mean=27 SD=5.99	mean=40 SD=10.28	mean=21.57 SD=4.49
BMI (Kg/m)	mean=10.24 SD=1.98	mean=15.89 SD=3.31	mean=9.11 SD=1.21
duration of disease	mean=3 year SD=1.58	mean=3.1 SD=1.8	—
daily insulin U/day	39.1 SD=7.6	mean=52.93 SD=17.03	—

none

Conclusion: the chronic increase in serum insulin level in insulin dependant diabetes mellitus children may be the cause of elevated serum leptin in these children. not only BMI but also insulin wee considered as significant pridictors of serum leptin cocentration. insulin, therefor, might plays a major. role inserum leptin concentration independant of obesity.

Reference(S): 1-Frederich R.c, Hamman A., Andrsn S., LollmanB., LowellBB., Flier JS.leptin levels reflect body lipid content in mice. evedence for diet induced resistance to leptin action. nature Medicine. 1995;1,1311.

2-mauzaki H, Ogawa Y , Isse N et al. Humman obese gene expression. Diabetes, 1995;44:855.

3-camfield LA., SmithFJ., Gusez Y., et al. Recombinant mouse ob protein: evidence for a propheral signal linking adiposity toand central neural networks. science. 1995;269:546.

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P1194

DIAGNOSIS AND MANAGEMENT OF BILIARY ATRESIA: THE KAOHSIUNG EXPERIENCE

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Introduction: To have a better understanding the character and the current management of the biliary atresia.

Methods: One hundred and thirty one patients were reviewed from charts with the diagnosis of biliary atresia. One hundred and thirty cases, sixty-six males and sixty-five females, were included.

Results: GGT/GOT was more than 2 in 78.5%, GGT/GPT in 83.4%, while 100% cases had alkaline phosphatase/GPT more than 2. Type I in 13 cases (9.9%), type II in 10 cases (7.6%), type III in 108 cases (82.4%), no expired cases were found in type I and type II. Choledochal cyst was found in 10 cases (7.6%). Combined with CMV infection was in 7 cases (5.3%), urinary tract infection diagnosed in 23 cases (17.5%, E. coli in 11). Sixty-five cases received portoenterostomy before 60 days old, 9 (13.8%) was jaundice free. Portoenterostomy after 60 days old was 4 cases (7.8%) jaundice free, after 90 days old there were no cases jaundice-free. Transplantation was indicated in the cases due to the characterization of worsening cholestasis, which led to portal hypertension and a decline in hepatic synthetic function. The age of receiving transplantation was from 6-month-20-day to 9-year-4-month old. The 1-year survival rate in portoenterostomy was 85.2%, 2-year 80%. Living donor liver transplantation 1-year survival rate was 97.1%, 2- years 96.4%.

Conclusion: Urinary tract infection should be checked in biliary atresia cases. Portoenterostomy, after 90 days old, has a poor jaundice free outcome. Liver transplanation, in the pediatric population, especially living related offers improved survival and quality of life.

Reference(S): 1.Howard ER, Tan KC: Br J Hosp Med 41:123, 1989

2. Martinez-Ibanez V, Bois-Ochoa J, Lloret J, et al: J Pediatr Surg 27:830, 1992

3. Vacanti J, Shamberger R, Erakalis A, et al: J Pediatr Surg 25:149, 1990.

4. Mckiernan PJ, Baker AJ, Kelly DA: Lancet 355:25, 2000

5. Silveira TR, Salzano FM, Howard ER, et al: Acta Paediatr Scand 80:1192, 1996

6. Vo Thi Diem H, Evrard V, Tran Vinh H, et al: Transplantation 75:1692, 2003

7. Al-Bassas A: Saudi Med J 24:S55, 2003

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P1195

ADVERSE EFFECTS OF FASTING THE PRETERM

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Introduction: This article discusses the controversy of fluid restriction and fasting of newborn & preterm versus fluid replenishment and early feeding which has persisted even to the present time. Thus, a brief history for this controversy is presented.

Methods: It has been found, through my work and observations in Neonatal Intensive Care Unit (NICU) over the last 20 years, that fasting the preterm led to many adverse effects. They have been divided into:

Results: A) Immediate Effects (or in the first 3 days):

(I) HYPOGLYCEMIA

(II) DEHYDRATION & electrolyte imbalance

(III) HYPERBILIRUBINEMIA

(IV) HYPOCALCEMIA & occasional associated HYPOMAGNESEMIA

B) Late adverse effects:

1) HYPERBILIRUBINEMIA

2) HYPOPROTEINEMIA

3) LOSS OF FAT

4) LOSS OF CALORIES („CHO)

5) HYPOVITAMINOSIS

6) COMPLICATIONS OF TOTAL PARENTERAL NUTRITION (TPN)

7) ANEMIAS OF PREMATURITY

During their discussion; my experiences, efforts and guide lines in the management of (NICU) have been illustrated also.

Conclusion: Early feeding of the preterm - either by fluids or milk - is advised to avoid all the above adverse effects.

Early enteral feeding - preferably by breast milk if possible - is better than parenteral feeding, but guided by the general condition of the baby and the occurrence of necrotising enterocolitis.

If breast milk suckling or expressed breast milk are not available or allowed, early preterm milk formula is advised.

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P1196

PANCREATITIS IN CHILDREN: A 10-YEAR ANALYSIS

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Introduction: Acute pancreatitis runs an unpredictable courses. The etiologies, clinical presentations and outcomes are different in children and adults. However, there has no reliable prognostic factor been reported in children's pancreatitis.

Methods: The medical records of 75 patients admitted due to pancreatitis from July 1992 to June 2002 were reviewed retrospectively. The diagnosis of pancreatitis was based on clinical symptoms, elevation of at least threefold of normal upper limit of pancreatic enzymes, and image findings. Data analyses were performed employing the Fisher's exact test for nominal variables, and nonparametric test for continuous variables.

Results: This study comprised 75 patients, including 37 males and 38 females, with 96 episodes of pancreatitis. The mean age was 10.1 ± 4.6 years (range 2 to 18 years). There were 20 patients with unknown etiology (26.7%); 15 with biliary disease (20.0%); 13 with abdominal trauma (17.3%).

The common clinical symptoms included abdominal pain (93.8%), vomiting (64.2%), and fever (33.3%). 55 patients were

managed conservatively, while the other 20 patients (26.7%) received surgical or other interventional treatment. 13 of them (65%) were associated with biliary tract diseases, 4 (20%) with abdominal trauma, and 3 (15%) with peritonitis.

In 85 episodes of non-traumatic pancreatitis, 17 episodes (20%) were severe pancreatitis. Among these patients, 12 had organ failure, DIC or metabolic disturbances, and the other 5 had local complications. 4 patients died of the disease. The mortality rate was 5.3%.

We compared initial clinical symptoms, laboratory parameters, and CT severity index between severe and mild pancreatitis. In severe patients, plasma AST, BUN, LDH and CRP level increased, while albumin and calcium levels decreased significantly. The Balthazar CT severity index (CTSI) >5 significantly correlated with severe pancreatitis ($p=0.03$), and CTSI>6 significantly correlated with death ($p=0.03$).

Conclusion: The overall outcomes of children's pancreatitis are good, but some of them developed severe disease and may require surgical intervention. CTSI is an applicable and comparable predictor for severe outcomes in children's pancreatitis.

Reference(S): 1. Yeung CY, et al. Pancreatitis in children: experience with 43 cases. *Eur J Pediatr*1996; 155:458-463

2. Simchuk, et al. Computed Tomography Severity Index Is a Predictor of Outcomes for Severe Pancreatitis. *Am J Surg* 2000; 179:352-5

3. Chatzicostas, et al. Balthazar Computed Tomography Severity Index Is Superior to Ranson Criteria and APACHE II and III Scoring Systems in Predicting Acute Pancreatitis Outcome. *J Clin Gastroenterol* 2003; 36:253-260

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P1197

IMMUNE RESPONSES IN VITRO TO THE HLA CLASS I-RESTRICTED A-GLIADIN PEPTIDE 123-132 (PEP A2) IN CULTURES OF INTESTINAL MUCOSA FROM COELIAC DISEASE PATIENTS ON GLUTEN-FREE DIET

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Introduction: Coeliac disease (CD) is associated with HLA class II-restricted CD4+ T mediated gliadin-specific responses, albeit a massive infiltration by CD8+ T cells represents the disease hallmark. A 10mer gliadin peptide (A-gliadin 123-132) is recognized by class I-restricted CD8+ T lymphocytes from both peripheral blood and intestinal mucosa of HLA-A2+ CD patients (1). We have tested in the organ culture of treated coeliac mucosa the capacity of A-gliadin 123-132 peptide to activate the local immune response.

Methods: Jejunal biopsies were obtained from 11 treated coeliac patients (6 HLA-A2 positive and 5 HLA-A2 negative), and in vitro cultured for 24 hours with A-gliadin 123-132 QLPCMD-VVL (pep A2). A peptic-tryptic digest of gliadin (PT-gliadin), or the immunodominant, A2-restricted, HIVgag 17 peptide SLYNTVATL (pep C), were used as controls. Immunohistochemical analysis was carried out assessing lamina propria CD25+ cells, IEL CD3+ cells density and FAS epithelial expression. Percentage of CD8/CD25, CD8/CD69, CD8/FAS-L double positive cells were evaluated by FACS analysis on cells isolated from two different CD biopsies cultured with medium or pep A2.

Results: In HLA A2+ coeliacs lamina propria CD25+ cells in biopsies cultured in the presence of pep A2 (median, range: 95, 38–107) or PT (122, 58–213), were significantly higher ($p < 0.05$) than in biopsies cultured with medium alone (27, 18–48) or pep C (27, 15–39). Similarly, the density of IEL CD3+/mm epithelium was higher for pep A2 and PT (42, 27–48; and 50, 39–71 vs medium: 28, 18–55 or pep C: 26, 23–28). Increased FAS epithelial expression was also noted. No differences were observed with pep A2 compared to medium in cultures of HLA A2- biopsies. An enhanced frequency of CD8/CD25 and CD8/FAS-L double positive cells was observed in cell preparations from pep A2-cultured biopsies compared to medium-cultured biopsies.

Conclusion: These data show that pep A2 is able to activate a peptide specific-immune response in HLA-A2+ coeliac patients and suggest the involvement of mucosa infiltrating CD8 T lymphocytes in CD pathogenesis. This work underlines also the importance of in vitro organ culture as a valuable tool to test the toxicity of gliadin peptides for coeliac patients.

Reference(S): 1. Gianfrani et al J Immunol. 2003;170:2719).

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P1198

WILSON'S DISEASE IN CHILDHOOD - PRESENTATION AND MANAGEMENT (4 YEARS RETROSPECTIVE STUDY)

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Introduction: Objective of the study: to evaluate the effectiveness of different treatments methods with chelating agents, Zinc or Orthotopic Liver Transplantation, based on hepatic or neurological manifestation at the pediatric patients with Wilson's Disease.

Methods: Between January 2000 and December 2003 14 children were Wilson's Disease diagnosed: 10 male and 4 female, with an average age of 10,2 years. The average duration from the beginning to the diagnosis was 14 months, longer at those ones with initial neurological disorder (20 months). The diagnosis was based on the low serum ceruloplasmin level (under 0,2g/l), increased of the copper in 24-hours urine and the presence of Kayser-Fleischer rings. The chelating drugs D-Penicillamine was the main stay of therapy for 10 patients (2 of them in association with Zinc); only 1 patient was treated with Zinc salts, one with Trientine and 2 benefited by Orthotopic Liver Transplantation. No severe side effects were noticed. We have regularly monitories (every 6 months): serum copper, ceruloplasmin, liver biochemistries and physical examination. We measured the copper excretion in 24-hours urine (under 0,5mg/l) every 6 months.

Results: All patients had a hepatic disease at the beginning of the study: hepatic cirrhosis (6 cases), chronic hepatitis (3), fulminate hepatic failure (3). 5 patients had a neuropsychiatric disorder. 2 patients with fulminate hepatic failure and hemolytic anemia who benefited by Orthotopic Liver Transplantation had a good further evolution. A patient with fulminate hepatic failure was treated with Trientine during a month, without improvement and evolution to exitus - we suspect poor compliance at the treatment. At the 5 patients with hepatic and neurological manifestation a partial remission of the neurological symptoms was achieved and the hepatic disease was stabilized. The neurological improvement was faster at those who received Zinc as well. At the 6 patients with hepatic disease, treated with D-Penicillamine a hepatic function improvement was noticed (3 by those ones were at presymptomatic stage).

Conclusion: 1. The liver disease (unknown origin) with positive family history should imply Wilson's Disease strongly.

2. The faster the introduction of the chelating therapy is, the better the results are.

3. At the symptomatic patients with a good compliance at the treatment an improvement of the hepatic disease was obtained.

4. The patients at whom the chelating treatment was initiated before the beginning of the clinical symptoms had an excellent evolution: no appearance of the symptoms on the followed period.

5. For patients with fulminate hepatic failure and hemolytic anemia the only solution was the Orthotopic Liver Transplantation.

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P1199

RECRUITMENT AND ACTIVATION OF DENDRITIC CELLS BY HELICOBACTER PYLORI

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Introduction: The means by which lymphocytes become activated in the gastric mucosa during *H. pylori* infection have not been described. Several different antigen presenting cell (APC) types may play a role in T cell activation including dendritic cells, macrophages, and epithelial cells. The aim of this study was to characterize and compare the phenotypes of these APCs following in vitro stimulation with *H. pylori*.

Methods: For immunofluorescence, frozen sections of gastric tissue were prepared from infected and naïve mice, and stained with labeled antibody specific for cell markers, and DAPI. The murine gastric epithelial cell line GSM06, freshly isolated peritoneal murine macrophages, and murine bone marrow derived dendritic cells were tested for activation in vitro. Dendritic cells were derived by culture of freshly isolated bone marrow cells in the presence of 10 ng/ml GM-CSF for 5 days, and then purified by positive selection for CD11c-positive cells. Purified dendritic, or the other cell types listed above, were then incubated for 48 hours in the presence of *H. pylori* SS bacteria, *H. pylori* antigen, or without stimulation. Supernatants were removed for cytokine analysis by multianalyte profiling and ELISA, and the cells were prepared for flow cytometry analysis using antibodies specific for CD80, CD86 or MHC Class II (I-A^b).

Results: CD11c-positive dendritic cells were prevalent in the gastric mucosa of infected mice compared to naïve mice. In culture, stimulation of dendritic cells resulted in morphologic changes including the development of dendrites. Stimulation of GSM06 cells resulted in marginal increases of inflammatory mediators but both macrophages and dendritic cells produced significantly higher amounts of IL-1², TNF α , and MIP-2, with the greatest increases observed for KC and IL-6. Flow cytometry analysis demonstrated that unstimulated dendritic cells displayed a higher percentage of positive cells for CD80, CD86 and MHC II than unstimulated macrophages. Stimulation with *H. pylori* resulted in increased, and ultimately equivalent expression of CD86 and MHC II on both dendritic cells and macrophages. However, CD80 expression of macrophages remained below 10%, even after stimulation, whereas dendritic cells increased expression 14% following stimulation to 46% positive cells.

Conclusion: Dendritic cells are present in the Helicobacter-infected gastric mucosa, and responsive to *H. pylori* stimulation in vitro. T cell stimulatory markers and cytokine expression indicate that dendritic cells may be a primary APC for immune induction during *H. pylori* infection.

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P1200

EFFICACY OF COMBINATION PROPHYLAXIS WITH SHORT-TERM HBIG AND LONG-TERM LAMIVUDINE IN PREVENTING DE NOVO HEPATITIS B INFECTION IN CHILDREN

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Introduction: We performed current study to evaluate the incidence of de novo HBV infection after liver transplantation according to the donor status of HBcAb and the efficacy of prophylaxis.

Methods: Non-concurrent comparative study was done in 61 children who received liver transplantation at Asan Medical Center from 1994 to 2001. We compared the incidence of de novo HBV infection according to the status of the donor HBcAb and analyzed the efficacy of prophylaxis. Lamivudine was administered at a dose of 2 mg/kg indefinitely. HBIG was administered under a standard protocol (100 IU/kg qd x7 days, then keep the titer of HBsAb > 100 mIU/mL for the first 6 months then stopped).

Results: All of 61 donors and recipients were HBsAg and HBV DNA negative preoperatively. Twenty seven (44%) of 61 were HBcAb positive and 17 of them received prophylaxis. While de novo HBV infection did not occur in 34 children with HBcAb negative donor, those in 10 children with HBcAb positive donor without prophylaxis developed in 4 children (40%). De novo HBV infection was not occurred among 17 children with prophylaxis. Outcomes of 4 children with de novo HBV infection with lamivudine therapy were disappearance of HBsAg in 1, negative sero-conversion of HBeAg in 1, and persistence of HBeAg in 2 children respectively.

Conclusion: HBcAb positive donor is associated with de novo HBV infection. Short-term HBIG and long-term lamivudine prophylaxis suggests to be effective in preventing de novo HBV infection in HBcAb positive donor graft.

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P1201

CHOLESTEROL ESTERS DISEASE..2 CASES REPORT

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Introduction: Cholesterol esters disease (ENEC), is a lipid metabolism autosomic recessive disease, due to acid lipase lysosomal (ALL) deficiency, a glycoprotein which hydrolyzes cholesterol esters and triglycerides in order to be used by cells; these molecules are deposited in different organs such as liver, spleen, small bowel, lymphatic nodules, suprarenal glands.

Methods: Case 1: 3 years old female. Who has initiated symptoms a year before with vomit, diarrhea, hyporexia, liver and spleen enlargement. Weight 11.6 kg, Height 82 cm. Liver was soft and smooth by palpation to 8 cm below costal edge, by total percussion 12 cm which cross medium line; spleen enlargement 4 cm. CBC: Hb 12.9, PMN 18,000, platelets 661,000. Glycemia 77, creatinine 0.3, AST 178, ALT 56, DHL 252, AP 271, GGT 62, TB 0.58, cholesterol 335, triglycerides 273. Ultrasonography study: liver and spleen enlargement with increase of echogenicity. Endoscopy: showed edema and the presence of velvet yellow appearance from the duodenal bulb up to third portion.

Duodenal biopsy: lamina propria partially infiltrated by granular and foamy macrophages. Liver biopsy: portal and pericellular fibrosis with granular and foamy macrophages. Case 2: 4 years old male. Who has initiated symptoms a year before with pale, vomit, weight loss, hyporexia, liver and spleen enlargement. Weight 13.5 kg, Height 94 cm. Liver was soft and smooth by palpation to 8 cm below costal edge, by total percussion 12.5 cm which cross medium line; spleen enlargement 4 cm. CBC: Hb 5.1, PMN 4,700, platelets 447,000. Glycemia 92, creatinine 0.25, AST 96, ALT 38, AP 158, GGT 59, cholesterol 266, triglycerides 258. Ultrasonography study: liver and spleen enlargement with normal echogenicity. Endoscopy: showed edema and the presence of velvet yellow appearance from the duodenal bulb up to third portion. Duodenal biopsy: lamina propria partially infiltrated by granular and foamy macrophages. Liver biopsy: small drops steatosis, portal and pericellular fibrosis with granular and foamy macrophages. Electronic microscopy: confirmed lipidic infiltration. Discussion: Both patients show by endoscopy lipidic deposits in small bowel; and the liver biopsy steatosis and granular and foamy macrophages. These findings confirmed the diagnosis.

Results: Both patients show by endoscopy lipidic deposits in small bowel; and the liver biopsy steatosis and granular and foamy macrophages. These findings confirmed the diagnosis.

Conclusion: Inborn errors of lipid metabolism should be consider in preschool children with hepato-splenomegaly and elevated serum cholesterol and triglycerides.

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P1202

THE PHARMACOKINETICS (PK) AND SAFETY OF A SINGLE DOSE OF ADEFOVIR DIPIVOXIL (ADV) IN CHILDREN AND ADOLESCENTS (AGED 2-17) WITH CHRONIC HEPATITIS B

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Introduction: ADV 10 mg is approved for the treatment of chronic hepatitis B (CHB) in adults. ADV as a 10 mg tablet and an oral suspension is in development for the treatment of CHB in pediatrics. The objective of this study was to evaluate the PK of adefovir in pediatrics and established dosing guidelines for patients aged 2-17 years.

Methods: Forty-seven HBeAg+ CHB patients with serum HBV DNA $\geq \log_{10}$ copies/mL (Roche Amplicor Monitor™, PCR), and ALT $\geq 2 \times \text{ULN}$ were enrolled in the study. Subjects 2-11 years received single doses of 0.14 mg/kg and 0.3 mg/kg (approximately $2 \times$ adult dose) in a cross over design with a 7-day washout period. Subjects 12-17 years received a single dose of 10 mg ADV. Following each dose, ADV levels were assessed at: 0, 0.5, 1, 2, 4, 6, 8, 12 and 24 hours post dose.

Results: Forty-five subjects were dosed. PK parameters of adefovir in subjects ≥ 12 years were similar to those seen in adults ($C_{\max} = 17.47 \text{ ng/mL}$ and $\text{AUC}_{\text{inf}} = 210.15 \text{ ng} \cdot \text{hr/mL}$). In subjects 2-6 and 7-11 years, a dose of 0.14 mg/kg resulted in relative underexposure; 0.3 mg/kg resulted in a higher C_{\max} but a comparable AUC_{inf} to adults. PK modeling was performed to identify a suitable dose for each age group. No clinically signifi-

cant adverse events were observed except one SAE unrelated to treatment.

Table 1: Age group and dose

		2-6 n=12	2-6 n=12	7-11 n=18	7-11 n=18	12-17 n=15
AUC inf.	[ng ² /mL]	0.14 mg/kg 104.74±33.76	0.3 mg/kg 224.13±78.72	0.14 mg/kg 128.48±53.81	0.3 mg/kg 292.44±105.08	10 mg 237.30±56.96
C _{max}	[ng/mL]	14.48±5.27	26.93±7.86	14.12±4.61	33.25±8.77	22.75±4.62
C _{last}	[ng/mL]	2.35±1.06	2.40±0.98	1.78±0.62	2.71±1.16	2.70±1.03

Conclusion: Based on this PK data, the proposed pediatric dose for phase 3 studies is 0.3 mg/kg for patients 2-6, 0.25 mg/kg for patients 7-11 and 10 mg for patients 12-17 years of age, with a maximum total dose of 10 mg daily, to achieve the target exposure or AUC of adefovir similar to the adult 10 mg dose. ADV was well tolerated at all doses.

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P1203

TREATMENT WITH THALIDOMIDE IN CHILDREN AND ADOLESCENT WITH INFLAMMATORY BOWEL DISEASE (IBD)

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Introduction: several small studies have evaluated the role of thalidomide in adults with refractory CD, but limited experience is available in pediatric population with IBD.

Aim: to assess the efficacy and the safety of low dose thalidomide in pediatric and adolescent patients with IBD, intolerant or refractory to conventional medical treatment.

Methods: we selected 23 patients (16 affected by CD and 7 by UC) steroid-dependent and refractory to either azathioprine or cyclosporine over 167 IBD patients (13.7%; CD, n=77; UC, n=69; indeterminate colitis, n=14) followed in our Pediatric Department. They received thalidomide 0.5-2 mg/kg/day orally and their response was assessed according to clinical score, colonoscopy, histological score and immunological methods.

Results: the mean age at IBD diagnosis was 10 y (range 1-17y). The mean duration of treatment with thalidomide was 20.6 months (range 1-63m). Eighteen over 23 patients (78%; CD=14/16, 87%; UC=4/7, 57%) had a significant clinical response, while 5/23 (3 with UC and 2 with CD) did not respond. The mean Pediatric Crohn Disease Activity Index decreased from 37.5 to 7. Steroid treatment was tapered and then discontinued in 16 patients within 1-3 months. Endoscopic and histological inflammation, C-protein and erythrocyte sedimentation rate (ERS) all decreased significantly. In one adolescent, thalidomide was effective in maintaining the infliximab-induced remission, allowing us to spare multiple infusions. Two patients affected by UC underwent surgery and stopped treatment. Side effects with the low dose thalidomide used were mild and mostly transient: sedation, dermatitis, drowsiness. Peripheral neuropathy occurred in 3 patients: the first one developed neuropathy after 5 years of treatment, then stopped thalidomide maintaining remission (follow-up after withdrawal of 2 years); the second one is still taking thalidomide because of good clinical response and the distal sensorial neuropathy is kept under control by administration of vitamin B12 and carnitine; the third one developed neuropathy after one month of treatment, then stopped thalidomide without any improvement of the intestinal symptoms.

Conclusion: our not controlled clinical study showed that thalidomide is well tolerated and effective in pediatric and adolescent IBD patients. Experiences with thalidomide in cases of refractory

IBD are very limited and the results of the first observational studies are very encouraging. Comparative studies are needed to define the therapeutic role of thalidomide in respect to other drugs such as infliximab, azathioprine and methotrexate

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P1204

TO BE ILL AS NARRATED BY CHILDREN 11-18 YEARS

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Introduction: The illness narrative can give professionals and caretakers a shared understanding of illness. The child's perspective of illness is seldom obtained. The aim with the study was to illuminate the experience of being temporarily ill at the age of 11-18 years.

Methods: Four girls and one boy, 11- 18 years old with temporarily illness were interviewed. They were acutely ill, injured, or underwent planned surgery. The interview was an open conversation and data was subjected to qualitative content analysis.

Results: To be ill was to be lost, lost in the body and lost in the feelings. To be ill was to be hurt, the body hurt and the children were hurt by others. To be ill was to be in need of comfort, comforting oneself and comforted by others.

Conclusion: To care for children at this age is to strike a balance between meeting dependency and independence. Routines and expectations can hinder nurses from being close to the children. The greatest task for professionals seems to be to meet the children with respect in their experience.

Reference(S): Burnard, P. (1991) A method of analysing interview transcripts in qualitative research. *Nurse Education today*, 11, 461-466

Burnard, P. (1996) Teaching the analysis of textual data: an experiential approach. *Nurse Education today*, 16; 278-281.

Riessman, C. K. (1993) *Narrative analysis*. Sage Publication, Newbury Park

Sörli, V., Jansson, L. & Norberg, A. (2003) The meaning of being in ethically difficult situations in paediatric care as narrated by female registered nurses. *Scandinavian Journal of Caring Science*, 17; 285-292.

Toombs, K.,S. (1992) The meaning of illness, A phenomenological account of the different perspective of physicians and patient. Kluwer Academic Publisher, Dordrecht

Woodgate, R. (2001) Adopting the qualitative paradigm to understanding children's perspectives of illness: barrier or facilitator? *Journal of Pediatric Nursing* 16; 149-161.

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P1205

RELATIONSHIP BETWEEN EARLY FEEDING AND COMMUNICATION DEVELOPMENT IN INFANTS: BIRTH TO 12 MONTHS.

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Introduction: Anecdotal reports suggest that infants experiencing early feeding difficulties may have delays in communication development

Methods: The first stage of the study involved the development of a theoretically based assessment protocol to test the

hypothesis of linked or independent processes required for feeding and speech. The assessment protocol will be piloted on 10 full term and 10 preterm infants at birth, 4, 8 and 12 months post term age. Measures of inter-judge and intra-judge reliability will also be taken of the observation protocol. Observation of the infant's language, speech, environment and maternal relationship will also be conducted to determine the nature and impact of environmental and social factors on feeding and/or speech and language development.

Results: To be analysed

Conclusion: It is hypothesised that infants experiencing early feeding difficulties and delay will also experience later speech and language delay.

Reference(S): Blanchard, I. (1963). "Better feeding can mean better speaking." *The American Journal of Nursing* 63(11).

Carpendale, J. I. and C. Lewis (in press). "Constructing an understanding of mind: the development of children's social understanding within social interaction." *Behavioral and Brain Sciences*.

Mathisen, B., L. Worrall, et al. (2000). "Feeding problems and dysphagia in six-month-old extremely low birth weight infants." *Advances in Speech-Language Pathology* 2(1): 9-17.

Starr, S. (2003). Facilitating early feeding and speech development. *Speech Pathology Australia*, Northam.

Ziegler, W. (2003). "Speech motor control is task-specific: Evidence from dysarthria and apraxia of speech." *Aphasiology* 17(1): 3-36.

Ziegler, W. (2003). "To speak or not to speak: Distinctions between speech and nonspeech motor control." *Aphasiology* 17(2): 99-105.

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P1206*

DEPRESSION OF WORKING NURSES' IN EDUCATIONAL UNIVERSITY HOSPITALS COMPARED WITH NURSING EDUCATORS IN NURSING & MIDWIFERY SCHOOL OF ISFAHAN UNIVERSITY OF MEDICAL SCIENCES

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Introduction: Nursing is an occupation taking care of humans' health. Nurses are targeted for depression due to numerous reasons as high stress existing in hospitals, critical status of the patients in pain arousing nurses' sympathy and higher number of female nurses compared with male nurses. Since nursing educators are in interaction with patients from one hand and with patients and students from other hand and as depression can be transferred from one to one, this study has tried to investigate average depression as the most indirect and hardest one is all above three groups.

Methods: This is a descriptive analytic cross sectional study. Beck depression test was used to collect depression. The population studied was composed of all nursing personnel including staff nurses and nurse educators working in Isfahan medical university hospitals with at least six months of work experience. The samples were selected by quota sampling. The collected data were analyzed by SPSS software with K- square test.

Results: The population studied included 395 working nurses in hospitals and 64 nurse educators working in nursing & Midwifery school with mean age of 33.47±0.4. There was no significant difference between the level of depression among nurse educators and working staff in hospitals there was more depression mentioned among single ones, females, staff on rotating morning evening shifts, subjects aged between 26 to 34 years and 1 to 5 years of experience, subjects with low job satisfaction, low social class of the job, words such as operating room, cancer

and critical units and low income.

There was a significant association between depression level and alternatives as working in shift, job satisfaction and the type of word.

Conclusion: The following items look to promote the mood of nurses if followed precisely:

- 1-Promotion of economic status
- 2-Disappearing the gap between theoretic and practical nursing
- 3-Providing hospitals with better facilities and technology
- 4-Making a more realistic suicide attitude toward nursing
- 5-Making physicians and other health staff acquainted with the role of nursing
- 6-On time encouragement and introducing out standing wards.
- 7-Making variation in working shifts for ward with higher risk of depression
- 8-Allocating more days of off and better facilities for rest time.
- 9-Specifying hospital green space for personnel usage
- 10-Respecting the items of risk factors mentioned in findings

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P1207

HELICOBACTER INFECTION IN CHILDREN

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Helicobacter Pylori (HP) in children causes some gastrointestinal diseases i.e chronic gastritis or peptic ulcer. The symptoms are Recurrent Abdominal Pain (RAP), dyspepsia, vomiting, hematemesis or anorexia. Most of our patients from previous study, the cause of these symptoms is HP. Therefore, HP diagnosis in children is important to make as early as possible and some investigators suggested to treat the patients especially the patients with symptoms.

The aim of our study is to know the incidence of HP in our populations, to report the symptoms of HP infection, the diagnosis of HP by endoscopic biopsy and the effect of HP treatment on the patient's symptoms.

Methodes: All patients with upper endoscopy included to this study except patients with esophageal varices or corpus alienum.

Result: From July 2002 to November 2003, 62 patient were included to the study. The average age is 5 years and 11 months (range from 1 months to 18 years). We found HP in 30 patients our of 62 patients (48%). The symptoms are RAP in 19 out of 30 patients (63.3%), dyspepsia in 5 patients (16.7%), hematemesis in 3 patients (10%) and anorexia in 3 patients (10%). Endoscopic diagnoses are Gastroduodenitis in 12 patients, Esofagogastro-duodenitis 5 patients, Esofagogastritis 5 patients and normal in 8 patients. Nodularity of duodenum were found in 8 patients (26.7%). From histopathologic examination, we found HP at duodenum in 21 patients, Anthrum pyloric in 24 patients and corpus in 9 patients. All patients were treated with triple therapy (Amoxycillin, clarithromycine and omeperazole) for 7 days. The symptoms were relieved in 27 patients (90%). Three patients still have RAP, which were cured after omeperazole administration for 2 weeks.

Conclusions:

1. HP incidence in our patients population is 48%
2. The most frequent symptom of HP infections in our patients is RAP
3. The other symptoms are hematemesis, dyspepsia and anorexia
4. We found HP more frequent at anthrum pylori compared to

duodenum and corpus gaster, but in 2 patients we only found HP at corpus

5. The treatment was succeeding to relieved the symptoms in most patients

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P1208

BILIARY ATRESIA IN INFANTS

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Kasai operation is still the firstline treatment for biliary atresia at our hospital. The aim of this study is to evaluate the outcome of infants with Biliary atresia.

Method: Retrospective analysis of medical records of patients with biliary atresia from 1999 to 2002

Result: Sixteen patients were included to the study. The average age of presentation was 2 months (range 1 month to 18 months). Two patients were not underwent Kasai procedure due to late diagnosis. Two patients were died immediately after the procedure and 1 patients after 10 months follow up. Kasai operation were performed in 9 patients during Golden age period. All patients had portoenterostomy done.

Complications included cholangitis (55%), Portal hipertension (75%) and hepatopulmonar syndrome (5%). The average time of jaundice free after operation was 5 month (2–12 months). Time for jaundice free was 3 months for patients who underwent Kasai during golden age period and 5 months after the period (statistically significant :p <0.05). The outcome of patients was related to clearance of jaundice (p <0.05).

Conclusions: We still had Biliary atresia patients after 3 months old at the first presentation. Kasai procedure helps patients but it was important to have this operation before 12 weeks old. Postoperative jaundice free can predict the outcome of our patients.

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P1210

THE STUDY OF LONG-TERM EFFECTS OF MARGINAL VITAMIN A DEFICIENCY DURING GESTATION AND TWO GENERATIONS ON GROWTH AND METABOLIC INDICES IN RAT.

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Background: Long-term effects of malnutrition on growth and metabolic indices are investigated in several case-control studies but the effects of long-term marginal vitamin A deficiency have not been studied yet.

Objective: In this regard a dietary study on growth and metabolic indices was conducted in a rat model during gestation and for two generations.

Methods: Firstly, seventeen pregnant rats were fed randomly a vitamin A sufficient (VAS) and vitamin A marginal (VAM) diet, from the first day of gestation until the last day of lactating their pups. In the next stage from female offspring in VAM, 14 female pups were fed randomly VAS (recovery group) or VAM diet. Seven female pups in VAS (control group) were fed the same diet. After mating and confirmation of pregnancy, feeding adult animals the same diet continued in each group till termination of lactation. This procedure continued until the puberty of second-

generation pups. Weight and height of all animals were recorded at puberty. Furthermore, hematological parameters (serum retinol, triglycerides, cholesterol and glucose) were studied in all animals after termination of lactating period. Obtained data were analyzed using ANOVA, Tukey, t-test, Mann-Whitney, Pearson Correlation and analysis of regression tests.

Results: Our results indicate that marginal vitamin A deficiency was accompanied with significant reduction in serum retinol in VAM group compared to VAS and recovery groups (P<0.001).

—Marginal vitamin A deficiency increased the serum triglyceride in VAM and recovery groups compared to VAS group significantly (P<0.01) and a negative significant correlation observed between retinol and serum triglyceride in the first (R=0.53, P<0.05) and the second generations (R=0.61, P<0.005).

—Marginal vitamin A deficiency increased the serum cholesterol from the zero to the second generations in VAM group significantly (P<0.001). Meanwhile serum cholesterol in recovery group was increased from the first to the second generations (P<0.01). And a significant quadratic correlation observed between retinol and serum cholesterol in the second generation (R=0.77, P<0.001).

—Marginal vitamin A deficiency increased the serum glucose in VAM group from the zero to the second generations (P<0.001). Meanwhile serum glucose in VAM and recovery groups were increased significantly compared to VAS group (P<0.005) and a negative significant correlation observed between retinol and serum glucose in the first (R=0.66, P<0.005) and the second generations (R=0.74, P<0.0001)

Conclusion: These data suggest that chronic marginal vitamin A deficiency deteriorate growth and metabolic indices in rat and these detrimental effects will be intensified when they are transferred to the next generations.

Key words: Vitamin A deficiency, gestation, growth index, metabolic indices, and rat

P1211

SURGICAL INTERVENTION IN INFLAMMATORY BOWEL DISEASE (IBD)

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As the incidence of IBD in children increases so does the requirement of surgical intervention. This presentation will give a brief overview of surgery in IBD. This will cover the different procedures in Crohn's disease and how this is related to the site affected (isolated small bowel disease, ileocaecal disease, colonic and anal disease), colectomy and either ileorectal anastomosis or ileal pouch anal anastomosis (IPAA) in ulcerative colitis and the problems that occur with these procedures. The presentation will cover the problems of IPAA procedure when the diagnosis is either 'indeterminate' or Crohn's colitis. Finally, the presentation will cover the use and problems with stoma's (ileostomy or colostomy) in children with IBD.

References: Keighley MRB and Williams NS. Surgery of the anus, rectum and colon. 2nd edition. 1999. WB Saunders London UK (This is the authoritative textbook on surgery in IBD)

Stringer MD et al. Paediatric surgery and urology: long term outcomes. 1st edition.

WB Saunders London UK Chapter 27 (This gives the surgery and outcome from the child's perspective)

P1212

NUTRITIONAL MANAGEMENT OF FAILURE TO THRIVE (FTT) IN DEVELOPING COUNTRIES: SOUTH AFRICA & AFRICA:

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Background: The treatment of FTT in SA and Africa, from its mildest form, malnutrition, to its most severe form, Kwashiorkor & Marasmus, is part & parcel of most health professional's day.

Poverty and HIV/AIDS significantly adds to the dilemma. The prevalence of HIV world wide is 41 million ('01); 28,1 million in Sub-Saharan Africa ('01), and 5 million in SA. That is 25% of the SA population, with \pm 250 000 <15 yrs old ('01').

Despite the fact that South Africa is relatively wealthy in comparison to the rest of Africa, a prevalence of 50% of the total SA population are classified as: Poor (Survive on <R1500/mnth (\pm 150 Euro) for a family of 2 adults & 2 children).

The SAVACG (95') study found that 23% (1 in 4) children in SA were stunted (reflection of SES), 9% (1 in 10) children in SA were underweight, 33% (1 in 3) children had marginal Vitamin A status, and 1 in 5 children was anaemic.

All stats included, are the latest trustworthy figures available.

Classification:

1. Malnutrition-FTT: \downarrow in wt centiles \geq 2 consecutive months, No gain in ht \geq 2 consecutive months (same wt) and a plato in weight / height gain \geq 2 consecutive months

2. Marasmus: According to the 'Wellcome Trust Classification (WTC): <60% expected wt for age, with no oedema present.

Clinical: Severe PEM, extreme weight loss, weakness.

3. Kwashiorkor: According to 'WTC: 60–80% of expected wt for age, with oedema present. **Clinical:** Hypoalbuminemia, Anaemia, Oedema, Pot belly, Depigmented skin, Hair is soft-brittle-sparse-reddish colour, undigested food in stools, liver is often fatty infiltrated

4. Marasmic-Kwashiorkor: According to 'WTC: <60% expected wt for age with / without oedema present.

Treatment: There is a SA Protein-Energy-Malnutrition Supplementation Programme 'NSP' that is operating from a community-clinic basis. Specific entry – and exit criteria has been laid down re. above discussed conditions, as well as for HIV/AIDS and breastfeeding mothers. The scheme provide an infant formula Nan-Pelargon, enriched porridge and a High Energy Drink, 1 yr +.

An important aspect of consulting with these cases, is also that of disease detection. An initial referral of malnutrition could more than often include HIV, TB, parasites / worms, FAS and mild to severe anaemia.

A very important aspect to keep in consideration when treating the cases mentioned in 1–4, is the dangers of the Refeeding Syndrome. During starvation fat and especially muscle stores (incl. the heart muscle) is the main source of fuel due to severe catabolism. During refeeding, carbohydrates become the main fuel source, which results in Insulin release, and enhanced uptake of Glucose, PO₄, K and water into cells. This could lead to GIT, Cardiopulmonary and Renal complications. Ileus, villi atrophy, polyuria, arrhythmias, Congestive Heart Failure and death, are some of these complications.

Conclusion: The treatment of malnutrition, poverty, TB and HIV/AIDS are continual elaborate challenges for the government and health professionals in SA and Africa. How do we solve the problem? *Treat them one by one – 'Mother Theresa.'*

References: 1. Eley BS, Sive A, Abelse L. Growth and micronutrient disturbances in stable HIV-infected children in Cape Town. *Ann of Tropical Paed* 2002; 22, 19–23

2. Solomon SM, Kirby DF. The Refeeding Syndrome: A Review. *JPEN* 1990; (14) 1, 90–97

P1213

Nutritional Treatment of Bilairy Atresia Patients:Pre-& Post Liver Transplant

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Background: Red Cross Children's Hospital (RXH), Cape Town, South Africa is the only dedicated children's hospital in SubSaharan Africa. It is also the only unit in Africa that does children's liver transplants between birth & 13 yrs old. Limited resources and organ donors, is the main reason why RXH has only performed 80 livertransplants over the last 20 years. These patients where mainly diagnosed with Bilairy Atresia and Hepatitis A. Living-related transplants only started in 2002. However, RXH remains a bright light of hope for Africa and SA.

Treatment:

1. Infant & Toddler Feeding: Breast is Best (lipase, DHA, AA etc.)

• Breast is Best! (Lipase, DHA, EPA, AA etc.).

Energy - 160 kcal/kg/day, Glucose polymer - \uparrow in 2% increments as tolerated until requirements are reached. MCT oil - \uparrow in 0.5% increments, as tolerated until requirements are reached. **Fat:CHO** ratio 40:60. **Protein** 4g/kg/day or [12–15% of total energy]. Modular/Hydrolysed Whey Protein - \uparrow in increments maintaining total protein ratio at 9 - 12% of total energy. **Volume:** 150 – 180 ml/kg

• **Alternative feeding:** Polymeric Infant feed with MCT's [palatable, available, cost], or, Semi-elemental Infant formulae

• **Optimal Neurological & Visual Development:** Formulae should contain / or be supplemented with: Docsa-hexaenoic Acid (DHA), Eicosapentaenoic acid (EPA) & Arachidonic Acid (AA)

• **BCAA:** Controversial: Preferentially used in catabolism to spare bodily prot. stores. Goal: Provide enough prot., without further liver deterioration & \downarrow Risk of encephalopathy (protect brain).

• **Weaning:** As any other infant only supplements could also now be added to the food.

• **Vitamins & Minerals:** Monitor Serum levels regularly (not always practical).

a. **Vitamin K:** On diagnosis – 1 mg IV, no improvement, continued & gradually changed to oral prep: 5–10 mg tablets wkly

b. **Vitamin D:** On diagnosis, 30 000 - 60 000i.u. repeated monthly for 3 months

c. **Vitamin E:** A single IM dose of 10mg/kg post opp - then 15mg - 200mg/kg/day orally

d. **Iron:** Therapeutic dose of oral iron, when deficiency is seen.

e. **Zinc:** 1mg/kg/d

2. The older child:

• From 10/12 to 1 yr +: Breast & or/ Polymeric Enteral Feed with MCT's

• Consider: Over night Pump-infused NG feeds

• Supplementation continues: MCT's, Protein, Glucose polymer

• **Energy:** 120–150% of RDA, **Protein:** 12–15 % of total energy

3. Nutritional Assessment:

• Serial measurements, Weight (Poor marker, oedema), Length, Head circumference, Skinfold thicknesses: supra-ileac often too painful / impossible to do accurately, Abdominal girth, Biochemical markers

4. Ascites:

• Fluid restriction, often: 80 – 100 ml/kg, Fully supplemented feeds: 1.6 – 2 kcal/ml

5. Oesophageal Varices:

• Banding / Sclerotherapy: Clear fluids 12 – 24 hrs post Rx, Soft diet 1wk +

6. Hepatic encephalopathy:

• Prot. restriction relative to degree of encephalopathy

- Grade 3/4 encephalopathy: NG feed, enough energy, minimum protein 0.5g/kg, intake in 0.5g/kg increments as improve

7. Liver Transplant:

- Fat Free TPN: First day/ 2, rarely more
- Start enteral feeding NGT / Oral a.s.a.p., usually day 2–3
- Extubated, stable: Diet appropriate for age
- F/up: Maintain good nutritional status

Conclusion: We thank a dedicated team of health professionals for fighting to maintain the only dedicated paediatric liver transplant unit in Sub-Saharan Africa.

References:

1. Otte JB. History of pediatric liver transplantation. Where are we coming from? Where do we stand? *Pediatr Transplant*. 2002 Oct; 6 (5): 378–87
2. The Liver and Pancreas. In: *Clinical Paediatrics Dietetics*. (Ed) Shaw V, Lawson M. Blackwell Publishing, 2001; 110–136.

ANSWERS/DISCUSSIONS

Clinical Quiz, continued from page 211

Answer: The patient underwent oesophagectomy with gastric interposition and pyloromyotomy. The biopsy result confirmed the diagnosis of oesophageal leiomyomatosis with irregular fragments of smooth muscle with associated fibrous tissue, blood vessels and nerve trunks. Currently, she is completely asymptomatic and is on full enteral feeds. She is gaining weight satisfactorily and is happy with her life.

Leiomyomatosis of the oesophagus is extremely rare condition in children (1). It should be distinguished from oesophageal leiomyoma, which is a localised capsulated benign lesion. Only 33 paediatric cases less than 18 years of age (including our case) have been reported in the literature (2–6). Clinical manifestations include vomiting, regurgitation, dysphagia, anorexia, cough, dyspnoea and/or cyanosis when swallowing, choking, hiccups, recurrent pneumonia, stridor and apnoea, faltering growth and weight loss and retrosternal pain. Haematemesis can occasionally occur as a clinical expression of oesophagitis (1–6). Other manifestations like haematuria and constipation are usually related to its association with Alport syndrome and leiomyomatosis at other sites (3–4).

Hall published the first report of a presumable oesophageal leiomyomatosis in 1916 (7). In 1983, Garcia-Torres and Gaurner first described an association of oesophageal leiomyomatosis and haematuric glomerular nephritis as a distinct clinical variant of Alport syndrome (haematuria, nephritis, sensorineural deafness and congenital cataract) (8). Since then, a number of cases have been reported, and a new term was coined: Alport Leiomyomatosis syndrome (1).

The diagnostic work up includes a detailed history and examination. Chest x-ray usually shows a wide mediastinum or a right-sided mediastinal mass. Diagnosis by barium studies and/or endoscopy is difficult as overlying mucosa remains intact. A thoracic CT or MRI scan is helpful but not diagnostic (5). Similarly, oesophageal manometry is also of little help in confirming the diagnosis. Endoscopic ultrasonography is described as a useful investigation by some authors (5).

The treatment is surgical. The affected oesophagus and gastric fundus, when involved, are resected and replaced with a gastric tube or colonic segment (1–6). Usually, a pyloromyotomy is also performed due to its association with pyloric stenosis. Outcome is good with disappearance of symptoms following surgery. No cases of relapse have been described until now in children after resection of the affected oesophagus for diffuse leiomyomatosis (1–6).

REFERENCES

1. Federici S, Ceccarelli PL, Bernardi F, et al. Esophageal leiomyomatosis in children: report of a case and review of the literature. *Eur J Pediatr Surg* 1998;8:358–63.
2. Lee H, Morgan K, Abramowsky C, Ricketts RR. Leiomyoma at the site of esophageal atresia repair. *J Pediatr Surg* 2001;36:1832–3.
3. Lerone M, Doredo P, Romeo G, et al. Leiomyomatosis of oesophagus, congenital cataracts and haematuria: report of a case with rectal involvement. *Pediatr Radiol* 1991;21:578–9.
4. Lonsdale RN, Roberts PF, Vaughan R, Thiru S. Familial oesophageal leiomyomatosis and nephropathy. *Histopathology* 1992;20:127–33.
5. Massicot R, Aubert D, Mboyo A, Destuynder O, Queneau PE. Localised esophageal leiomyoma and hypertrophic osteoarthropathy. *J Pediatr Surg* 1997;32:646–7.
6. Bourque MD, Spigland N, Bensoussan AL, et al. Esophageal leiomyoma in children: two case reports and review of the literature. *J Pediatr Surg* 1989;24:1103–7.
7. Hall AJ. A case of diffuse fibromyoma of the oesophagus, causing dysphagia and death. *Quart J Med* 1916;9:409–28.
8. Garcia-Torres R, Gaurner V. Leiomyomatosis del esofago, traqueobronchial y genital asociada con nefropatia hereditaria tipo Alport: un nuevo sindrome. *Rev Gastroenterol Mex* 1983;48:163–70.